

Hypoplastic left heart syndrome with anomalous circumflex artery arising from left pulmonary artery. A description of combined surgical and interventional cardiac treatment

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Hypoplastic left heart syndrome (HLHS) belongs to the group of congenital complex heart defects that are characterized by hypoplasia of the left heart and the aorta [1]. The described coronary anomaly (anomalous left coronary artery arising from the pulmonary artery (ALCAPA)) is relatively rare in the group of patients with HLHS [1–4].

We present case of an HLHS patient, who was subjected to a hybrid procedure at 25 days old. In the sixth month of life, the patient was readmitted for planned reconstruction of the aorta, joining the aorta and the pulmonary artery trunk and performing the Glenn operation with pulmonary arterioplasty and left pulmonary artery (LPA) stent implantation (bare-metal stent, 10 × 17 mm). On the first postoperative day, the child's status was critical, with a markedly decreased right ventricular systolic function (EF = 38%) and intense generalized stress reaction. Urgent catheterization demonstrated the anomalous circumflex artery (Cx) arising from the distal segment of the LPA (ALCAPA, Figure 1 A). Catheterization showed the persistent left superior vena cava (L-SVC) draining directly into the coronary sinus which was embolized by a vascular Amplatzer plug (Figure 1 B).

A reoperation was necessary, consisting in grafting the Cx to the neo-aorta. In the postoperative period, the patient presented with numerous episodes of cardiac rhythm disturbances (supraventricular tachycardia). In view of the persisting low saturation values, i.e. oxygen saturation (O₂) of 60%, arterial partial pressure of oxygen (PaO₂) of 28–30 mm Hg, and distal LPA narrowing (Figure 1 C), another catheterization was achieved on the 10th day after surgery, during which LPA stent redilatation was performed (Figure 1 D).

After the procedure, oxygenation improved (sat. O₂ 85%; PaO₂ 38–40 mm Hg) and the heart rhythm disturbances gradually resolved. The child was discharged home 14 days after stent implantation. At present, the 12-month old child is in a good general condition, presenting no signs of overt heart insufficiency. Her saturation is maintained at O₂ 80% and EF in echocardiography is 50%.

The available literature presents only a few descriptions of patients with HLHS and ALCAPA; in the vast majority of cases, such patients did not live beyond the first postoperative day [1, 2].

The first stage of HLHS treatment is a severe trauma for the child, and the surgery itself, as well as the duration of cardiopulmonary bypass, may be long [3]. Therefore, the probability of the patient developing coronary ischemia increases, especially in subjects with anomalies of the coronary arteries [4]. Thanks to interdisciplinary collaboration of cardiac surgeons, interventional cardiologists and echocardiographers, our patient successfully completed her surgical treatment and was discharged; at home, she is currently awaiting the next stage of treatment (Fontan procedure).

Coronary anomalies in the form of abnormal origin of the coronary vessels are very rare abnormalities in HLHS, and such abnormalities continue to pose a challenge for a cardiac surgical team. Careful observation of the patient after the first stage of surgery and before qualification for the next stage allows alarming episodes, e.g. desaturation, to be noted, in which case the diagnostic management should be extended.

Conflict of interest

The authors declare no conflict of interest.

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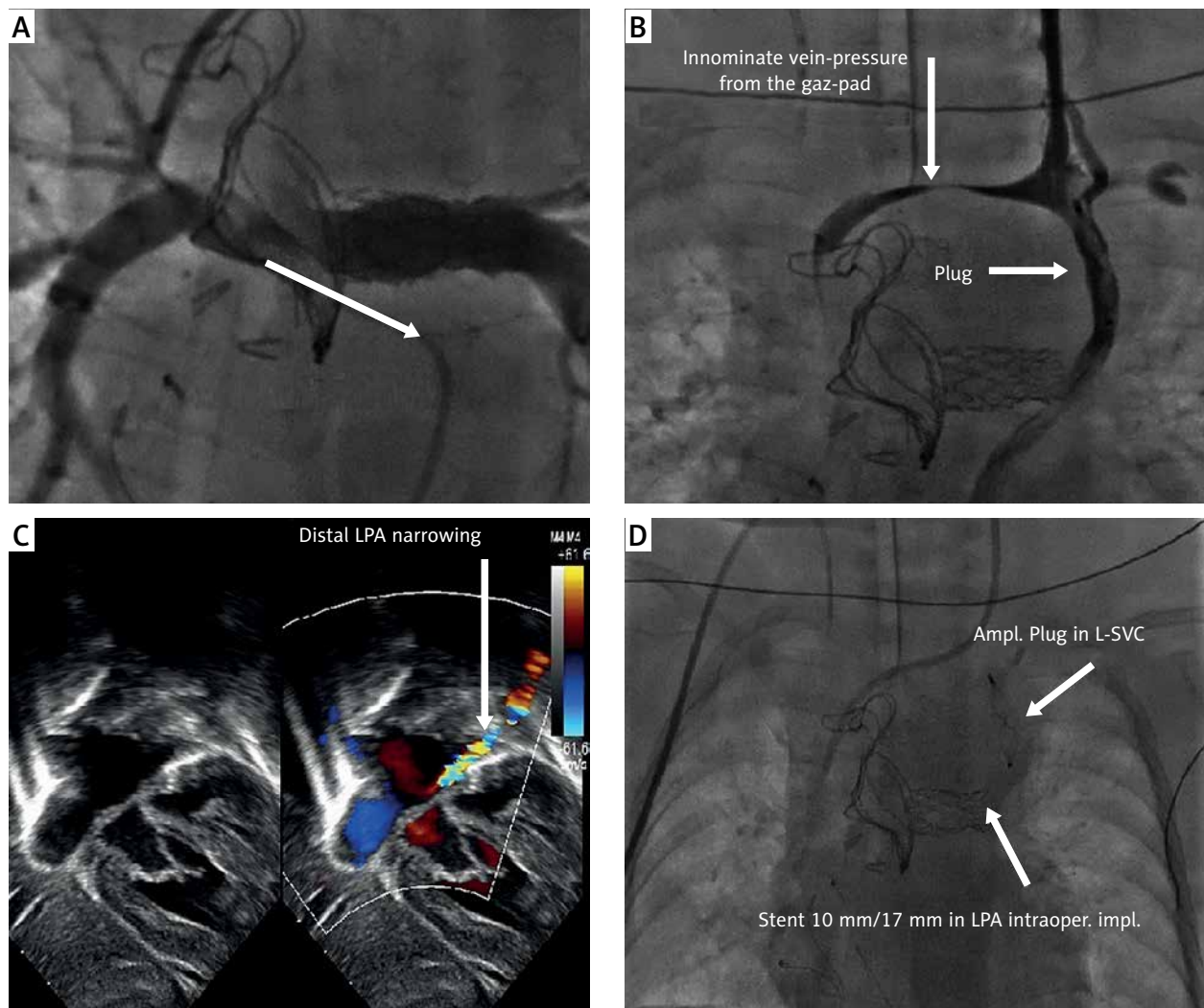


Figure 1. **A** – Postoperative heart catheterization showing anomalous circumflex artery arising from the distal segment of the left pulmonary artery. **B** – Postoperative heart catheterization demonstrated persistent left superior vena cava (L-SVC) Heart team (cardiologist, cardiac surgeon, intervention cardiologist) decided to embolize by vascular Amplatzer plug implantation. Catheterization was performed early after surgery. The patient had been still on open chest due to gaz-pad being illustrated on the radiogram. **C** – Transthoracic echocardiography demonstrated distal left pulmonary artery (LPA) stent narrowing with Doppler gradient 60–70 mm Hg. **D** – Next catheterization procedures with LPA stent redilatation

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